Questions & Answers about . . .

Juvenile Rheumatoid Arthritis

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)
National Institutes of Health
Public Health Service • U.S. Department of Health and Human Services

For Your Information

This publication contains information about medications used to treat the health condition discussed in this booklet. When this booklet was printed, we included the most up-to-date (accurate) information available. Occasionally, new information on medication is released.

For updates and for any questions about any medications you are taking, please contact the U.S. Food and Drug Administration at 1–888–INFO–FDA (1–888–463–6332, a toll-free call) or visit their Web site at www.fda.gov.

Additional copies of this booklet are available from

National Institute of Arthritis and Musculoskeletal and Skin Diseases
NIAMS/National Institutes of Health
1 AMS Circle
Bethesda, MD 20892–3675

You can also find this booklet on the NIAMS Web site at www.niams.nih.gov/hi/topics/juvenile_arthritis/juvarthr.htm.

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What Is Arthritis?

Arthritis means "joint inflammation" and refers to a group of diseases that cause pain, swelling, stiffness, and loss of motion in the joints. "Arthritis" is often used as a more general term to refer to the more than 100 rheumatic diseases that may affect the joints but can also cause pain, swelling, and stiffness in other supporting structures of the body such as muscles, tendons, ligaments, and bones. Some rheumatic diseases can affect other parts of the body, including various internal organs. Children can develop almost all types of arthritis that affect adults, but the most common type that affects children is juvenile rheumatoid arthritis (JRA).

What Is Juvenile Rheumatoid Arthritis?

Juvenile rheumatoid arthritis is arthritis that causes joint inflammation and stiffness for more than 6 weeks in a child of 16 years of age or less. Inflammation causes redness, swelling, warmth, and soreness in the joints, although many children with JRA do not complain of joint pain. Any joint can be affected and inflammation may limit the mobility of affected joints. One type of JRA can also affect the internal organs. Doctors classify JRA into three types by the number of joints involved, the symptoms, and the presence or absence of certain antibodies found by a blood test. (Antibodies are special proteins made by the immune system.) These classifications help the doctor determine how the

disease will progress and whether the internal organs or skin is affected.

- **Pauciarticular** (PAW-see-are-TICK-you-lar)—Pauciarticular means that four or fewer joints are affected. Pauciarticular is the most common form of JRA; about half of all children with JRA have this type. Pauciarticular disease typically affects large joints, such as the knees. Girls under age 8 are most likely to develop this type of JRA.

  Some children have special kinds of antibodies in the blood. One is called antinuclear antibody (ANA) and one is called rheumatoid factor. Eye disease affects about 20 to 30 percent of children with pauciarticular JRA. Up to 80 percent of those with eye disease also test positive for ANA and the disease tends to develop at a particularly early age in these children. Regular examinations by an ophthalmologist (a doctor who specializes in eye diseases) are necessary to prevent serious eye problems such as iritis (inflammation of the iris, the colored part of the eye) or uveitis (inflammation of the uvea, or the inner eye). Some children with pauciarticular disease outgrow arthritis by adulthood, although eye problems can continue and joint symptoms may recur in some people.

- **Polyarticular**—About 30 percent of all children with JRA have polyarticular disease. In polyarticular dis-

ease, five or more joints are affected. The small joints, such as those in the hands and feet, are most commonly involved, but the disease may also affect large joints. Polyarticular JRA often is symmetrical; that is, it affects the same joint on both sides of the body. Some children with polyarticular disease have an antibody in their blood called IgM rheumatoid factor (RF). These children often have a more severe form of the disease, which doctors consider to be similar in many ways to adult rheumatoid arthritis.

- **Systemic**—Besides joint swelling, the systemic form of JRA is characterized by fever and a light skin rash, and may also affect internal organs such as the heart, liver, spleen, and lymph nodes. Doctors sometimes call it Still's disease. Almost all children with this type of JRA test negative for both RF and ANA. The systemic form affects 20 percent of all children with JRA. A small percentage of these children develop arthritis in many joints and can have severe arthritis that continues into adulthood.

What Causes Juvenile Rheumatoid Arthritis?

JRA is an autoimmune disorder, which means that the body mistakenly identifies some of its own cells and tissues as foreign. The immune system, which normally helps to fight off harmful, foreign substances such as bacteria or viruses, begins to attack healthy cells and tissues. The result is

inflammation—marked by redness, heat, pain, and swelling. Doctors do not know why the immune system goes awry in children who develop JRA. Scientists suspect that it is a two-step process. First, something in a child's genetic makeup gives them a tendency to develop JRA; then an environmental factor, such as a virus, triggers the development of JRA.

What Are the Symptoms and Signs of Juvenile Rheumatoid Arthritis?

The most common symptom of all types of JRA is persistent joint swelling, pain, and stiffness that typically is worse in the morning or after a nap. The pain may limit movement of the affected joint although many children, especially younger ones, will not complain of pain. JRA commonly affects the knees and joints in the hands and feet. One of the earliest signs of JRA may be limping in the morning because of an affected knee. Besides joint symptoms, children with systemic JRA have a high fever and a light skin rash. The rash and fever may appear and disappear very quickly. Systemic JRA also may cause the lymph nodes located in the neck and other parts of the body to swell. In some cases (less than half), internal organs including the heart and, very rarely, the lungs may be involved.

Eye inflammation is a potentially severe complication that sometimes occurs in children with pauciarticular JRA. Eye diseases such as iritis and uveitis often are not present until some time after a child first develops JRA.

Typically, there are periods when the symptoms of JRA are better or disappear (remissions) and times when symptoms are worse (flare-ups). JRA is different in each child—some may have just one or two flare-ups and never have symptoms again, while others experience many flare-ups or even have symptoms that never go away.

Some children with JRA may have growth problems. Depending on the severity of the disease and the joints involved, growth in affected joints may be too fast or too slow, causing one leg or arm to be longer than the other. Overall growth may also be slowed. Doctors are exploring the use of growth hormones to treat this problem. JRA also may cause joints to grow unevenly or to one side.

How Is Juvenile Rheumatoid Arthritis Diagnosed?

Doctors usually suspect JRA, along with several other possible conditions, when they see children with persistent joint pain or swelling, unexplained skin rashes and fever, or swelling of lymph nodes or inflammation of internal organs. A diagnosis of JRA also is considered in children with an unexplained limp or excessive clumsiness.

No one test can be used to diagnose JRA. A doctor diagnoses JRA by carefully examining the patient and considering the patient's medical history, the results of laboratory tests, and x rays that help rule out other conditions.

- **Symptoms**—One important consideration in diagnosing JRA is the length of time that symptoms have been present. Joint swelling or pain must last for at least 6 weeks for the doctor to consider a diagnosis of JRA. Because this factor is so important, it may be useful to keep a record of the symptoms, when they first appeared, and when they are worse or better.

- **Laboratory tests**—Laboratory tests, usually blood tests, cannot by themselves provide the doctor with a clear diagnosis. But these tests can be used to help rule out other conditions and to help classify the type of JRA that a patient has. Blood may be taken to test for RF and ANA, and to determine the erythrocyte sedimentation rate (ESR).
  - — ANA is found in the blood more often than RF, and both are found in only a small portion of JRA patients. The RF test helps the doctor tell the difference among the three types of JRA.
  - — ESR is a test that measures how quickly red blood cells fall to the bottom of a test tube. Some people with rheumatic disease have an elevated ESR or "sed rate" (cells fall quickly to the bottom of the test tube), showing that there is inflammation in the body. Not all children with active joint inflammation have an elevated ESR.

- **X rays**—X rays are needed if the doctor suspects injury to the bone or unusual bone development. Early in the disease, some x rays can show cartilage

damage. In general, x rays are more useful later in the disease, when bones may be affected.

- **Other diseases**—Because there are many causes of joint pain and swelling, the doctor must rule out other conditions before diagnosing JRA. These include physical injury, bacterial or viral infection, Lyme disease, inflammatory bowel disease, lupus, dermatomyositis, and some forms of cancer. The doctor may use additional laboratory tests to help rule out these and other possible conditions.

Who Treats Juvenile Rheumatoid Arthritis? What Are the Treatments?

The special expertise of rheumatologists in caring for patients with JRA is extremely valuable. Pediatric rheumatologists are trained in both pediatrics and rheumatology and are best equipped to deal with the complex problems of children with arthritis and other rheumatic diseases. However, there are very few such specialists, and some areas of the country have none at all. In such circumstances, a team approach involving the child's pediatrician and a rheumatologist with experience in both adult and pediatric rheumatic disease provides optimal care for children with arthritis. Other important members of the team include physical therapists and occupational therapists.

The main goals of treatment are to preserve a high level of physical and social functioning and maintain a good quality

of life. To achieve these goals, doctors recommend treatments to reduce swelling; maintain full movement in the affected joints; relieve pain; and identify, treat, and prevent complications. Most children with JRA need medication and physical therapy to reach these goals.

Several types of medication are available to treat JRA:

- **Nonsteroidal anti-inflammatory drugs (NSAIDs)**—Aspirin, ibuprofen (Motrin, Advil, Nuprin),* and naproxen or naproxen sodium (Naprosyn, Aleve) are examples of NSAIDs. They often are the first type of medication used. Most doctors do not treat children with aspirin because of the possibility that it will cause bleeding problems, stomach upset, liver problems, or Reye's syndrome. But for some children, aspirin in the correct dose (measured by blood test) can control JRA symptoms effectively with few serious side effects.

  If the doctor prefers not to use aspirin, other NSAIDs are available. For example, in addition to those mentioned above, diclofenac and tolmetin are available with a doctor's prescription. Studies show that these medications are as effective as aspirin with fewer side effects. An upset stomach is the most common complaint. Any side effects should be reported to the

* Brand names included in this booklet are provided as examples only, and their inclusion does not mean that these products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

doctor, who may change the type or amount of medication.

- **Disease-modifying anti-rheumatic drugs (DMARDs)**—If NSAIDs do not relieve symptoms of JRA, the doctor is likely to prescribe this type of medication. DMARDs slow the progression of JRA, but because they take weeks or months to relieve symptoms, they often are taken with an NSAID. Various types of DMARDs are available. Doctors are likely to use one type of DMARD, methotrexate, for children with JRA.

  Researchers have learned that methotrexate is safe and effective for some children with rheumatoid arthritis whose symptoms are not relieved by other medications. Because only small doses of methotrexate are needed to relieve arthritis symptoms, potentially dangerous side effects rarely occur. The most serious complication is liver damage, but it can be avoided with regular blood screening tests and doctor followup. Careful monitoring for side effects is important for people taking methotrexate. When side effects are noticed early, the doctor can reduce the dose and eliminate side effects.

- **Corticosteroids**—In children with very severe JRA, stronger medicines may be needed to stop serious symptoms such as inflammation of the sac around the heart (pericarditis). Corticosteroids like pred-

nisone may be added to the treatment plan to control severe symptoms. This medication can be given either intravenously (directly into the vein) or by mouth. Corticosteroids can interfere with a child's normal growth and can cause other side effects, such as a round face, weakened bones, and increased susceptibility to infections. Once the medication controls severe symptoms, the doctor may reduce the dose gradually and eventually stop it completely. Because it can be dangerous to stop taking corticosteroids suddenly, it is important that the patient carefully follow the doctor's instructions about how to take or reduce the dose.

- **Biologic agents**—Children with polyarticular JRA who have gotten little relief from other drugs may be given one of a new class of drug treatments called "biologic agents." Etanercept (Enbrel), for example, is such an agent. It blocks the actions of tumor necrosis factor, a naturally occurring protein in the body that helps cause inflammation.

- **Physical therapy**—Exercise is an important part of a child's treatment plan. It can help to maintain muscle tone and preserve and recover the range of motion of the joints. A physiatrist (rehabilitation specialist) or a physical therapist can design an appropriate exercise program for a person with JRA. The specialist also may recommend using splints and

other devices to help maintain normal bone and joint growth.

- **Complementary and alternative medicine**—Many adults seek alternative ways of treating arthritis, such as special diets or supplements. Although these methods may not be harmful in and of themselves, no research to date shows that they help. Some people have tried acupuncture, in which thin needles are inserted at specific points in the body. Others have tried glucosamine and chondroitin sulfate, two natural substances found in and around cartilage cells, for osteoarthritis of the knee.

  Some alternative or complementary approaches may help a child to cope with or reduce some of the stress of living with a chronic illness. If the doctor feels the approach has value and will not harm the child, it can be incorporated into the treatment plan. However, it is important not to neglect regular health care or treatment of serious symptoms.

How Can the Family Help a Child Live Well With JRA?

JRA affects the entire family who must cope with the special challenges of this disease. JRA can strain a child's participation in social and after-school activities and make school work more difficult. There are several things that family

members can do to help the child do well physically and emotionally.

- Treat the child as normally as possible.

- Ensure that the child receives appropriate medical care and follows the doctor's instructions. Many treatment options are available, and because JRA is different in each child, what works for one may not work for another. If the medications that the doctor prescribes do not relieve symptoms or if they cause unpleasant side effects, patients and parents should discuss other choices with their doctor. A person with JRA can be more active when symptoms are controlled.

- Encourage exercise and physical therapy for the child. For many young people, exercise and physical therapy play important roles in managing JRA. Parents can arrange for children to participate in activities that the doctor recommends. During symptom-free periods, many doctors suggest playing team sports or doing other activities to help keep the joints strong and flexible and to provide play time with other children and encourage appropriate social development.

- Work closely with the school to develop a suitable lesson plan for the child and to educate the teacher and the child's classmates about JRA. (See the end of this booklet for information about Kids on the Block, Inc., a program that uses puppets to illustrate how juvenile arthritis can affect school, sports, friends, and family.) Some children with JRA may be absent from school for prolonged periods and need to have the teacher send assignments home. Some minor changes such as an extra set of books, or leaving class a few minutes early to get to the next class on time can be a great help. With proper attention, most children progress normally through school.

- Explain to the child that getting JRA is nobody's fault. Some children believe that JRA is a punishment for something they did.

- Consider joining a support group. The American Juvenile Arthritis Organization runs support groups for people with JRA and their families. Support group meetings provide the chance to talk to other young people and parents of children with JRA and may help a child and the family cope with the condition.

- Work with therapists or social workers to adapt more easily to the lifestyle change JRA may bring.

Do Children With Juvenile Rheumatoid Arthritis Have To Limit Activities?

Although pain sometimes limits physical activity, exercise is important to reduce the symptoms of JRA and maintain function and range of motion of the joints. Most children with JRA can take part fully in physical activities and sports when their symptoms are under control. During a disease flare-up, however, the doctor may advise limiting certain activities depending on the joints involved. Once the flare-up is over, a child can start regular activities again.

Swimming is particularly useful because it uses many joints and muscles without putting weight on the joints. A doctor or physical therapist can recommend exercises and activities.

What Are Researchers Trying To Learn About Juvenile Rheumatoid Arthritis?

Scientists are investigating the possible causes of JRA. Researchers suspect that both genetic and environmental factors are involved in development of the disease and they are studying these factors in detail. To help explore the role of genetics, the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) has established a research registry for families in which two or more siblings have JRA. NIAMS also funds a Multipurpose Arthritis and Musculoskeletal Diseases Center (MAMDC) that specializes in research on pediatric rheumatic diseases including JRA.

The research registry for JRA is located at Children's Hospital Medical Center at the University of Cincinnati College of Medicine in Ohio. The registry, established in 1994, continues to list new cases as well as be maintained and systematically updated. The focus of the registry is on families whose brothers and sisters have JRA, with emphasis on genetic susceptibility in those affected families.

Researchers are continuing to try to improve existing treatments and find new medicines that will work better with fewer side effects. For example, researchers are studying the long-term effects of the use of methotrexate in children. In addition, the Food and Drug Administration's "Pediatric Rule" requires manufacturers of new drugs and biologic agents, such as etanercept, that will be commonly used for children to provide specific information about safe pediatric use.

Where Can People Get More Information About the MAMDC?

For more information about the MAMDC, contact:

David N. Glass, M.D.
Children's Hospital Medical Center
3333 Burnet Avenue
Cincinnati, OH 45229–3039
Phone: 513–636–7686 (administrative office),
513–636–4676 (clinic)
E-mail: david.glass@chmcc.org
www.cincinnatichildrens.org/Research/Divisions/Rheumatology/default.htm

Where Can People Get More Information About Juvenile Rheumatoid Arthritis?

- **National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)**
  National Institutes of Health
  1 AMS Circle
  Bethesda, MD 20892–3675
  Phone: 301–495–4484 or
  877–22–NIAMS (226–4267) (free of charge)
  TTY: 301–565–2966
  Fax: 301–718–6366
  E-mail: NIAMSInfo@mail.nih.gov
  www.niams.nih.gov

  NIAMS provides information about various forms of arthritis and rheumatic disease and bone, muscle, joint, and skin diseases. It distributes patient and professional education materials and refers people to other sources of information. Additional information and updates can also be found on the NIAMS Web site.

- **American Academy of Orthopaedic Surgeons**
  P.O. Box 2058
  Des Plaines, IL 60017
  Phone: 800–824–BONE (2663) (free of charge)
  www.aaos.org

  The academy provides education and practice management services for orthopaedic surgeons and allied health professionals. It also serves as an advocate for improved

patient care and informs the public about the science of orthopaedics. The orthopaedist's scope of practice includes disorders of the body's bones, joints, ligaments, muscles, and tendons. For a single copy of an AAOS brochure, send a self-addressed stamped envelope to the address above or visit the AAOS Web site.

- **American College of Rheumatology**
  1800 Century Place, Suite 250
  Atlanta, GA 30345
  Phone: 404–633–3777
  Fax: 404–633–1870
  www.rheumatology.org

  The association provides referrals to doctors and health professionals who work on arthritis, rheumatic diseases, and related conditions. The association also provides educational materials and guidelines.

- **American Juvenile Arthritis Organization**
  1330 West Peachtree Street
  Atlanta, GA 30309
  Phone: 404–872–7100 or 800–283–7800 (free of charge)
  www.arthritis.org

  Part of the National Arthritis Foundation, this organization is the primary nonprofit group devoted to childhood rheumatic diseases. It has information about JRA, support groups, and pediatric rheumatology centers around the country.

- **Kids on the Block, Inc.**
  9385–C Gerwig Lane
  Columbia, MD 21046
  Phone: 410–290–9095 or
  800–368–KIDS (5437) (free of charge)

  Kids on the Block, Inc., is an educational program that uses puppets to show how JRA can affect school, sports, friends, and family. A package is available (for a fee) that includes a set of large puppets that represent a diverse group of children, as well as audiocassettes, a training guide, four different program scripts, props, followup activities, and other resources. The program is designed so that anyone can be a puppeteer, and workshops to train puppeteers are available.

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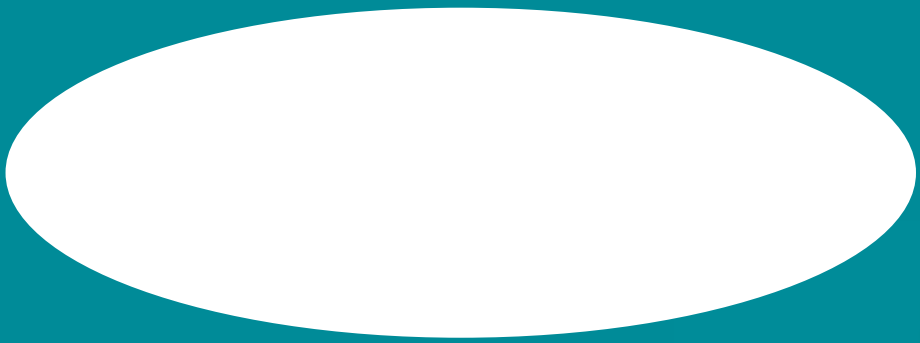
The mission of the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), a part of the National Institutes of Health (NIH), is to support research into the causes, treatment, and prevention of arthritis and musculoskeletal and skin diseases, the training of basic and clinical scientists to carry out this research, and the dissemination of information on research progress in these diseases. The National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse is a public service sponsored by the NIAMS that provides health information and information sources. Additional information can be found on the NIAMS Web site at www.niams.nih.gov.



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